WS12.5 Lung clearance index and exercise capacity among children with mild CF- and non-CF bronchiectasis

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Objectives: To compare FEV_1, LCI and exercise capacity among children with CF and non-CF bronchiectasis and healthy controls.
Methods: Eighteen stable children and adolescents with CF, 15 stable children with non-CF bronchiectasis and 20 healthy children and adolescents were requited. Spirometry and multiple-breath-washout tests were performed. All children performed a cardiopulmonary exercise test on cycle ergometer. Children with CF and non-CF bronchiectasis had a HRCT scan, which was evaluated using the modified Bhalla score.
Results: The patients' mean age was 14.7 years, mean FEV_1 75.8% predicted, while the healthy children's mean age was 14.9 years, mean FEV_1 98.9%. Total Bhala score and FEV_1 did not differ significantly among children with CF and non-CF bronchiectasis ($p > 0.05$). Mean VOpeak was significantly impaired among children with bronchiectasis compared to healthy children ($p < 0.01$), but did not differ significantly among children with CF and non-CF bronchiectasis (p: 0.225). Patients with CF bronhiectasis had significantly higher respiratory equivalent to carbon dioxide (VE/VCO2) and higher physiologic dead space to tidal volume ratio (VD/VT), during exercise, compared to non-CF patients ($p < 0.001$). LCI was significantly increased among patients with bronchiectasis compared to healthy children ($p < 0.01$). LCI did not differ significantly among children with CF and non-CF bronchiectasis (p: 0.22).
Conclusions: Exercise testing and Multiple Breath Washout measurements can discriminate children with mild bronchiecatsis from healthy children. However, the burden of the disease is more prominent in children with CF compared with the non-CF bronchiectasis.

WS12.6 Comparison of two methods to identify bronchiectasis on chest computed tomography scans in two cohorts of young patients with cystic fibrosis

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Background: Bronchiectasis (BE) detected by chest CT in young children is an important outcome measure for clinical studies. For these studies it is important to use objective, sensitive and reproducible methods to detect and monitor progression of BE.
Aim: To compare sensitivity of two methods to detect BE on chest CTs in young CF patients.
Methods: Routine chest CTs of 10 patients aged 2 years were randomly selected from the Sophia Children's Hospital and of 10 patients aged 3 years from the Princess Margaret Hospital for Children. In addition the follow up chest CTs of these children 2 years later (age 4 and 5 years) were selected. CTs were randomized and de-identified (Myrian, Intrasense, France). BE was scored using the CF-CT scoring module in all scans. In the same batch, all airway-artery (AA) pairs visible on the CTs were numbered and scored for each generation as normal or as BE where the AA-ratio was >1.
Results: No significant differences in incidence of BE were observed between the 2 cohorts studied. From *CF-CT scoring:* 1st CT, BE was present in 11 patients (55%), compared to 13 patients (65%) at follow up CT. *AA > 1:* 1st CT BE was present in 3 patients (15%), compared to 10 patients (50%) at the follow up CT.
Conclusion: This study supports the progressive nature of BE and prevalence of BE in young children, as observed in previous studies. The AA-ratio method underestimates the prevalence of BE relative to CF-CT scoring method in children age 2–3 years. We speculate that CF-CT scoring more readily detects abnormal airways which are often not accompanied by visible arteries and thus not included in the AA analysis.

WS12.7 Pulmonary emphysema in cystic fibrosis detected by densitometry on chest multidetector computed tomography (MDCT)

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Background: Early histopathological studies on lung specimens from patients with cystic fibrosis (CF) and recent results from a mouse model indicate that emphysema may contribute to CF lung disease. However, little is known about the relevance of emphysema in patients with CF. In the present study, we used computationally generated density masks based on multidetector computed tomography (MDCT) of the chest for non-invasive characterization and quantification of emphysema in CF.
Methods: Volumetric MDCT scans were acquired in parallel to pulmonary function testing in 41 patients with CF and 21 non-CF controls, and subjected to dedicated software. The lung was segmented, low attenuation volumes below a threshold of –950 Hounsfield units were assigned to emphysema volume (EV), and the emphysema index (EI) was computed. Results were correlated with forced expiratory volume in 1 s percent predicted (FEV1%), residual volume (RV), and RV/total lung capacity (RV/TLC).
Results: We show that EV was increased in CF (457±530 ml) compared to non-CF controls (78±90 ml) ($P < 0.01$). EI was also increased in CF (7.7±7.5%) compared to the control group (1.2±1.4%) ($P < 0.05$). EI correlated inversely with FEV1% ($r_s = -0.66$), and directly with RV ($r_s = 0.69$) and RV/TLC ($r_s = 0.47$) in patients with CF ($P < 0.007$), but not in non-CF controls. Emphysema in CF was detected from early adolescence (~13 years) and increased with age ($r_s = 0.67$, $P < 0.001$).
Conclusions: Our results indicate that early onset emphysema detected by densitometry on chest MDCT is a characteristic pathology that contributes to airflow limitation and may serve as a novel therapeutic target in advanced CF.